1: Int J Dermatol. 1998 Mar;37(3):211-4.

Familial pemphigus vulgaris in mother and daughter.

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BACKGROUND: Pemphigus vulgaris is an autoimmune disease in which genetics appears to be of basic importance. Although association with certain human leukocyte antigen (HLA) alleles has been found in some ethnic groups and individuals, no true disease susceptibility genes have been established, and familial cases are very unusual. METHODS: We report a Polish family with pemphigus vulgaris in the mother and daughter. The diagnosis was confirmed by cytologic, histologic, and immunofluorescence studies. RESULTS: The course was severe and the disease long-lasting in the mother, probably due to treatment with small doses of corticosteroids without immunosuppressive drugs. In the daughter, treated with larger doses of corticosteroids and azathioprine, the lesions regressed within 4 months, after which maintenance therapy was instituted with 10 mg of prednisone daily. The HLA studies performed in the daughter and her three children after the mother had died showed identical haplotypes in both the patient and the healthy children. The patient has given birth to a healthy child while still having a high titer of intercellular (IC) antibodies. CONCLUSIONS: The familial occurrence of pemphigus in first-degree relatives is suggestive of inherited susceptibility to the disease, transmitted as a dominant trait. The identical haplotypes in the healthy children of the patient favor the role of other, unknown factors required for the development of the disease in predisposed individuals.

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- Case Reports

MeSH Terms:
- Adult
- Aged
- Diagnosis, Differential

- Female
- HLA Antigens/genetics
- Haplotypes
- Humans
- Pemphigus, Benign Familial/diagnosis*
- Pemphigus, Benign Familial/drug therapy
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